SHORT REVIEW

The acute idiopathic scrotal edema; a diagnostic challenge

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ABSTRACT: The acute idiopathic scrotal edema (AISE) is a self-limited disease of unknown etiology, characterized by edema and erythema of the scrotum and the dartos, without expanding to the underlying layers of scrotum's wall or to the endoscrotal structures. About 60 to 90% of all cases involve boys younger than 10 years old. Based on the classic references, diagnosis is made after excluding the main causes of painful scrotum, especially those that require urgent intervention. Thorough clinical examination conducted by the pediatric surgeon in conjunction with the pathognomonic findings of color flow Doppler ultrasound, especially the fountain sign, eliminate significantly the need for urgent surgical intervention of the scrotum, due to diagnostic doubts. This review article aims at presenting the most recent data about the AISE, after systematic consultation of the international references.

Keywords: Painful scrotum, acute idiopathic scrotal edema (AISE), fountain sign, male child

INTRODUCTION

The term "acute idiopathic scrotal edema" (AISE) was first used by Qvist in 1956 (1). It was the announcement of a study including 158 infants and young boys suffering from acute scrotal edema, which resolved in a few days, without complications. Of all the diseases that come under the entity of painful scrotum, 2.3% of these cases are attributed to the AISE (2). This is a self-limited disease, characterized by swelling and erythema of the scrotum and the dartos, without expanding to the underlying layers of scrotal wall or the endoscrotal structures (3,4). It starts typically from the groin or the perineum, but rapidly expands to the scrotum (5).

A differentiation is determined concerning about the localization of AISE: Geiger J. Epelman M. and Darge K. (6) and Lee A., Park S., et al (7) in recent publications found that 75% of all cases were bilateral. Klin B., Lotan G., Efrati Y. et al (8) found that 90% of the cases were unilateral. The majority of AISE cases appear in patients aged 5-11 years old. 60 to 90% of all cases involve boys younger than 10 years old (6).

ETIOLOGY

The etiology of AISE is currently unknown, although certain situations have occasionally been implicated in the pathogenesis of this entity, such as:

- the cellulitis, that extends from the perianal region to the scrotum. This approach lacks, because, on the one hand, the cultures received from the region are usually negative, and on the other hand, the AISE is mainly self-limited without therapeutic intervention
- ii. the bites caused by insects
- iii. an injury
- iv. the urine loss, that causes aseptic inflammatory response in the area, expanding to the scrotum
- v. the allergy. It is considered as the most likely cause, based on the fact that eosinophilia is found in more than 40% of all AISE cases, while these patients suffer also from diseases such as asthma, eczema or contact dermatitis. Besides, there is evidence that medication with antihistamines contributes to the recession of the symptoms. Thus, it is believed that AISE is a clinical manifestation of angioedema. (9,10,11)

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CLINICAL FEATURES

These patients usually do not complain about accompanying symptoms. They can rarely mention indistinct discomfort in the scrotum. It is unusual, when itchiness precedes edema. Generally, scrotal edema is the first clinical manifestation, it develops suddenly and is accompanied with diffuse erythema in the affected area. AISE spreads rapidly, within a few hours. Swelling often begins unilaterally (Left side/Right side=1), but progressively extends to the whole scrotum. Erythema expands to the groin (67% of all cases), the perineum (42%), the penis (20%) or the suprapubic region. Edema and erythema are the main manifestations of this entity in all patients, while local pain coexists in 80% of AISE cases. The duration of the presence of the above clinical features is 6 to 72 hours (average 14 hours) (6). Soon begins the recession of edema and erythema, which is usually completed within 48 hours, without residual lesions. In 21% of all cases, AISE recurs from 1 to 3 times, but no complications arise. The time, that relapse occurs, ranges from several months to years.

DIAGNOSIS

Physical examination evinces that no systematic symptoms exist. Edema, erythema and possibly abolition of the normal ripples of scrotum due to the contraction of the dartos are noted. After thorough examination of the endoscrotal structures, no pathological signs arise. However, when scrotal edema is excessive, pediatric surgeon may encounter difficulty in the process of objective examination. During palpation of the scrotum, mild sensitivity and slight temperature rise are typical findings. If the surgeon exercises gentle pressure instantaneously, and then stops this manipulation, he will ascertain the formation of an imprint, which is indicative of vascular stasis. Also, the presence of swollen inguinal lymph nodes can be noted.

Based on international references, differential diagnosis includes causes of acute painful scrotum, such as the torsion of the spermatic cord, the torsion embryonic appendages, the epididymitis, an injury, the testicular or paratesticular tumors, allergic purpura, Fournier's gangrene and a strangulated inguinal hernia. The primary concern of the clinician is to exclude nosologic entities that require urgent surgical intervention, such as spermatic cord torsion.

Pathognomonic clinical signs of the testicular torsion are the absence of the cremasteric reflex and the higher

testicle's position. Pathognomonic clinical findings in patients suffering from appendiceal torsion are the localized tenderness, while palpating the upper pole of the testis, and the blue-dot sign, during transillumination of the affected hemiscrotum. Eventually, pathognomonic finding of the epididymitis is the sensitivity, while palpating the affected, apparently expanded epididymis.

Basic diagnostic tool in clinical practice is the color flow Doppler ultrasound, which contributes to the exclusion of other basic causes of acute painful scrotum, mainly those that require immediate surgical intervention, and the diagnostic documentation of the AISE, evincing the main, pathognomonic imaging findings.

The exclusion of the testicular torsion (TT) can be done in an accurate way, by conducting high resolution ultrasound imaging using frequencies 10-12 MHz: primary indication of TT is the appointment of rotations of the spermatic cord (snail-shell shaped mass), and not the disruption of the testicular circulation (6,12,13). In terms of sensitivity, high resolution ultrasound has 96% sensitivity, while color flow Doppler ultrasound 76%.

After performing cross sections, the thickening of scrotal wall is highlighted, ranging from 3.4 to 13.4 mm (average 7, 7 mm). This thickening is usually homogenous. A pathognomonic finding is the increased vascularity of the scrotal layers but with normal appearance of the testis and the epididymis. Blood supply of the scrotum is ensured by the anterior and posterior scrotal artery, which originate from deep branches of the internal and external pudendal artery. Imaging of these vascular branches in cross sections, along with the scrotal hyperemia, constitutes the characteristic fountain sign (6, 14). Other possible ultrasonographic findings are both the development of reactive hydrocele (20% of all cases) and the swollen and hyperemic inguinal lymph nodes (4).

TREATMENT

First of all, it is significant that the clinician reassures the parents about the disease's progression and of course about the non-development of complications. Usually, AISE is self-limited within the first 48-72 hours, thus, this time monitoring of the patient is required. Monitoring can also take place outpatient. Auxiliary interventions that can be implemented, however, without adequate substantiation, is the decline of physical activity and the raise of the scrotum. No references exist in order to prove the possible assistance by administering antimicrobial or anti-inflammatory drugs.

However, it is clear that, when the patient is given antihistamines, disease's recession is faster.

CONCLUSIONS

Previously, the diagnosis of AISE was made after excluding other causes of acute painful scrotum. Nowadays, thorough clinical examination by the pediatric surgeon in conjunction with the demonstration of pathognomonic findings after conducting ultrasonographic imaging, especially the fountain sign, reduces significantly the need for urgent surgical investigation of the scrotum, due to diagnostic doubts, especially with respect to the TT, when this entity cannot be excluded.

Το οξύ ιδιοπαθές οίδημα του οσχέου: μια διαγνωστική πρόκληση

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Περίληψη: Είναι αυτοπεριοριζόμενη νόσος με άγνωστη αιτιολογία χαρακτηριζόμενη από οίδημα και ερύθημα του οσχέου και του δαρτού χωρίς επέκταση στα υποκείμενα τοιχωματικά στρώματα του τοιχώματος ή στις ενδοοσχεικές δομές. Στο 60-90% των περιπτώσεων αφορά αγόρια ηλικίας κάτω των 10 ετών. Με βάση την κλασσική βιβλιογραφία η διάγνωση τίθεται αποκλείοντας τα αίτια του επώδυνου οσχέου, κυρίως αυτών που απαιτούν επείγουσα επέμβαση. Η ενδελεχής κλινική εξέταση από τον ειδικό χειρουργό παίδων σε συνδυασμό με την ανάδειξη των παθογνωμονικών ευρημάτων του Doppler έγχρωμης ροής και ιδιαίτερα του σημείου πίδακα, περιορίζει σημαντικά την αναγκαιότητα της επείγουσας διερεύνησης του οσχέου λόγω διαγνωστικής αμφιβολίας.

Σκοπός της παρούσας μελέτης είναι η παράθεση των πλέον πρόσφατων δεδομένων μέσα από την αναδίφηση της πρόσφατης βιβλιογραφίας.

Λέξεις Κλειδιά: Επώδυνο όσχεο, ιδιοπαθές οίδημα οσχέου, σημείο πίδακα, αγόρι

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